of cleavage has been located, that is, the line of cleavage between the two layers of the fascia of Denonvillier, the rectum strips back with the greatest of ease and the seminal vesicles are very quickly brought into view. I wish to state here that the vesicles are by no means in a dark field and when properly exposed should be almost on a level with the skin surface of the perineum. This is accomplished without any undue tension on the tractor. The fascia of Denonvillier is now opened on each side just over the seminal vesicles and they can either be opened up and cauterized with carbolic acid or can be picked up at their tips and easily removed. It should be remembered, however, that the ampulla of the vas presents small diverticula not far different from the diverticula of the seminal vesicles, and it is believed that in practically every case, whether the seminal vesicles are removed or not, the ampulla of the vas should be drained.

Bleeding following seminal vesiculectomy is only very slight and the shock of the operation so little as to be astonishing. As a rule the wound should be healed thoroughly within two weeks.

Some operators advise the insertion of a rubber tube into the rectum after the operation has been completed and the use of opium pills to prevent the bowels from moving for four or five days. From my experience with Young's perineal prostatectomy and with a limited number of seminal vesiculectomies I am thoroughly convinced that the rectal plug only causes discomfort to the patient and is more likely to produce a rectal fistula than not and should not be used. It has been our custom to give castor oil on the second day, as it adds a good deal to the post-operative comfort and convalescence.

The operation of seminal vesiculectomy is extremely simple but should not be undertaken unless one is thoroughly familiar with the technique of Young's perineal prostatectomy and with the anatomy of the perineum.

MONGOLISM.*

By RACHEL L. ASH, M. D., San Francisco.

The term *Mongolism* denotes a congenital state of mental deficiency, whose most marked characteristics are certain resemblances to the Mongol race.

The first definite description of this condition was given by Langdon-Down, in 1866; Shuttleworth added many important observations; while Telford-Smith, in 1896, developed mongolian imbecility into a clinical entity.

Mongolian imbeciles are found in all countries, and form from 3 to 5 per cent. of the feeble-minded in institutions. As the great majority of mongols die in early childhood, the actual number must be much greater, the institutional statistics representing merely the survivors.

PATHOLOGICAL ANATOMY.

The result of autopsies has been disappointing; more knowledge has been obtained by clinical study.

The Head. The head is distinctly micro-brachycephalic; the circumference is small; the vertex, low; the antero-posterior diameter is less than the transverse. Occasionally the shape is complicated by rickets, hydrocephalus, etc. The perpendicular frontal bone is parallel to the occiput. Usually the malar bones are prominent. The base of the skull is small; the pharyngeal vault is low and narrow, and the nasal cavities are diminished in size.

The Long Bones. The long bones show nothing characteristic except that they do not grow to the length of these bones in normal individuals. This accounts for the dwarfism of this type.

The Hand. The hand is the most remarkable feature of the mongol skeleton. The metacarpus is small; the metacarpal fingers are short and thick, but tapering toward the extremities. The thumb and little finger are very short. The little finger is incurved toward its neighbor, its tip seldom reaches to the middle of the second phalanx. It is known as the Telford-Smith finger. The X-ray shows a very small second phalanx, frequently with a change in the size and shape of the distal phalanx. The time of ossification is normal; occasionally, it may be premature.

The Brain. Much time has been consecrated to the study of the brain, but the results of all investigations may be summed up thus, to quote from Vogt: "While many cases reveal normal findings, in the majority of the cases are seen: lack of development of entire cerebral lobes, or portions of these, such as single convolutions, abnormal smallness of single lobes, the pons, the medulla, the cerebellum, and, lastly, delayed, undeveloped cerebral gray and white matter. All these point to an impeded progress in the last stage of foetal development."

Many changes have been observed in the ear, heart, and intestines, but these are common in other defectives and do not deserve special mention here.

No constant variation from the normal has been found in the anatomical study of the ductless glands.

CLINICAL FEATURES.

The appearance of the mongol is typical. The main features are noticeable at birth,—the small head, the slant eyes, the saddle nose, the thick, protruding tongue; the child nurses with difficulty. As he grows older, other characteristics make their appearance. Very characteristic is the lack of tone of the muscles, permitting the child to assume many unusual and normally impossible positions with its limbs. The hands with the short thickened fingers, the protruberent abdomen, the inability of the child to sit up, the late appearance of the teeth, the sucking tongue, the absent speech, and the lack of interest in its surroundings early call the attention of the mother to an unusual

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* Read before the Forty-sixth Annual Meeting of the Medical Society of the State of California, Coronado, April, 1917.

condition in her offspring. Since the capacity for inspired and expired air is diminished, the respirations are shallower and more frequent than normal. The area of mucous membrane is diminished, and, aided by the protruded tongue, it becomes insensitive; consequently, infections are frequent. Chronic coryza, bronchitis and blepharitis are the rule.

The hair is usually sparse and is always straight. There is no constant peculiarity in the shape of the ears. The face is striking. It is round and often flat; the cheeks are inclined to be chubby and are often flushed. The forehead is usually of good shape and is not as wrinkled as that of the cretin. The nose is short and flat; the bridge, broad and sunken; the overlying skin is lax and frequently extends over the eyes for some distance as epicanthic folds, thus giving an appearance of increased distance between the eyes. The eyes appear small owing to the narrowness of the palpebral fissure, which is still further characterized by an oblique slant which originally suggested the name *mongolism*. The mouth is small, and is usually kept open to protrude the large, coarse, dry and fissured tongue.

The skin is generally rough and dry, thickened and mottled. The forehead, cheeks, neck and back of hand often suggest myxoedema.

Secondary sexual characteristics develop late. Frequently the testes are undescended and the menses much delayed. Pregnancy in a mongol is unknown, although other classes of low-grade defectives have borne children.

The haemoglobin is normal, and the cellular elements present nothing unusual. The blood pressure is generally 50 to 80 mm., below the average for the age. Extensive work has been done on the metabolism of mongols. The sugar tolerance is high; the Ca and P output in the urine and faeces is much increased.

The mongol is always undersized. Even if at birth the child is of normal weight and length, after the third year the rate of growth diminishes. At ten years of age he is 2½ to 4½ inches shorter than normal. As is to be expected from the lack of tonicity of the muscles, the child balances himself with difficulty and begins to walk very late, as a rule between the third and fourth year.

In the nervous system, aside from a diminished sensation to external stimuli, heat and cold, touch and pain (very common in defectives), there is nothing of special interest. The tendon reflexes are present.

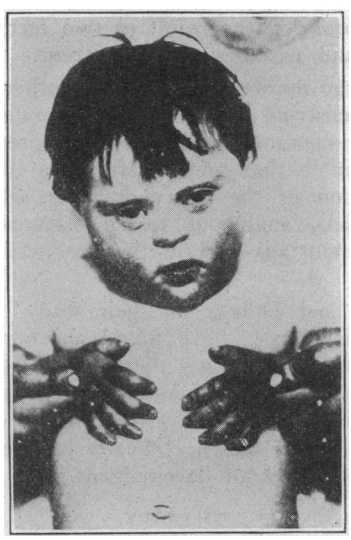

Fig. 1.
Mongolian Idiot; aet. four years. Note typical hands.

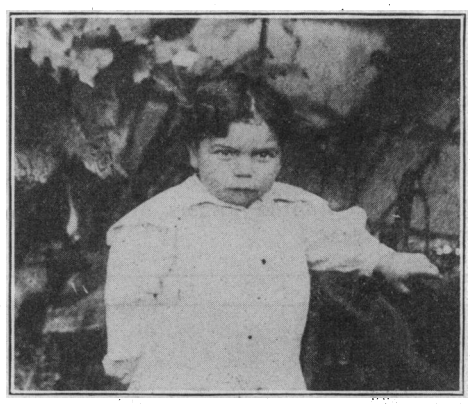

Fig. 2.
Cretin; aet. four years.

MENTALITY.

As the child grows older, his lack of mentality becomes more and more apparent. What in early childhood was thought to be obedience, is really an indifference to surroundings. With locomotion, the real character of the child becomes evident. Sometimes there are uncontrollable fits of anger, though as a rule the mongol imbecile is of a happy disposition. He is an excellent imitator and is even able to dance in time to music. The mentality, according to authorities, rarely reaches the sixth year, and is never beyond the eighth year, so that he is usually classed as a low-grade imbecile.

Speech is learned very late, and it is rare to hear fully developed sentences, even at mature age.

DIAGNOSIS.

The diagnosis of mongolism is easily made. Some difficulty may arise, however, in differentiating it from *cretinism*, and, in early life, from those diseases in which locomotion is delayed—rachitis, amyotonia congenita, and achondroplasia.

Cretinism. The characteristic appearance of the cretin is due to an absence or lack of function of the thyroid. His skin is myxoedematous. His skull is large; the frontal bosses, prominent and the occiput, conspicuous. The long bones are thick and short, the appearance of the centers of ossification exceedingly delayed. The hand is usually small and immature, the thumb and little finger are normal, but the last phalanges of all the fingers are short and broad. The malar bones are normal as are usually the eyes. There is no sad-

dle nose. The face is thick, pale, and with few lines of facial expression. The cretin is stupid, obstinate and bad-tempered. With him the thyroid therapy brings about happy results. His idiocy may be of extreme grade.

Rachitis. The broad head, the softness of the bony structures with consequent deformity, the changes in the epiphyses, and the unclouded mind serve to distinguish this condition.

Amyotonia congenita. There is the same hypermobility here as in mongolism, but the facies are of a different type, the tendon reflexes are absent and the mentality is normal for the age of the child.

Achondroplasia. Confusion will arise only in the first months of life; the characteristic large head, the long body, the short limbs, the trident hand with the blunted fingers are noticeable before the first year.

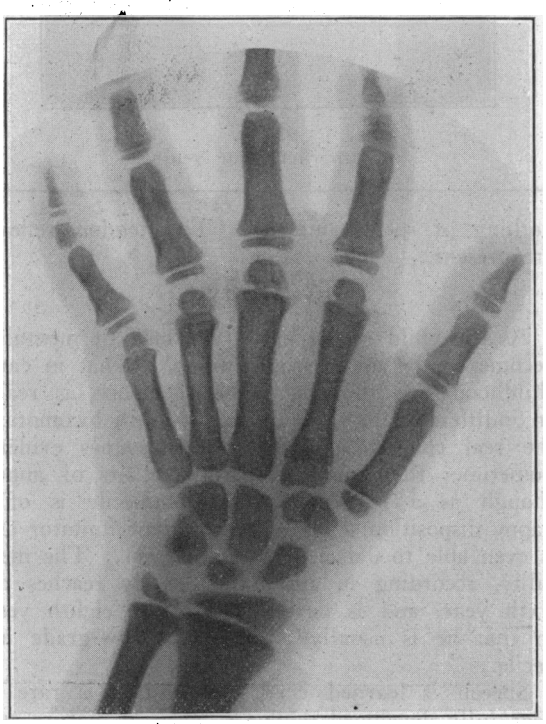

Fig. 3.
Radiogram of hand of Mongolian imbecile, age seven years. Note the Telford-Smith finger, and a typical ossification of the second phalanx of the index finger.

PROGNOSIS.

Mongols do not live very long. Forty years is a very unusual age, and the majority die in early childhood. More than 50 per cent. die of some infection of the respiratory tract; tuberculosis or pneumonia are the most frequent.

PATHOGENESIS.

Syphilis, tuberculosis, and alcoholism of the parent occur no more frequently in mongols than in other defectives.

From the figures given by different observers, it would seem that more than half of the mongols are the last-born children often of large families where the offspring have come in rapid succession; that one or both parents are in or beyond the third decade; and that the mother has been weakened by physical or mental strain. It may be added that, in some cases, the mothers are very young; here the reproductive powers may not yet be fully established. Which of these conditions is the most potent is yet in doubt. Probably all act concurrently in many cases. Mongolism is an exhaustion product; the result of two factors, morbid heredity and weakness of the parent.

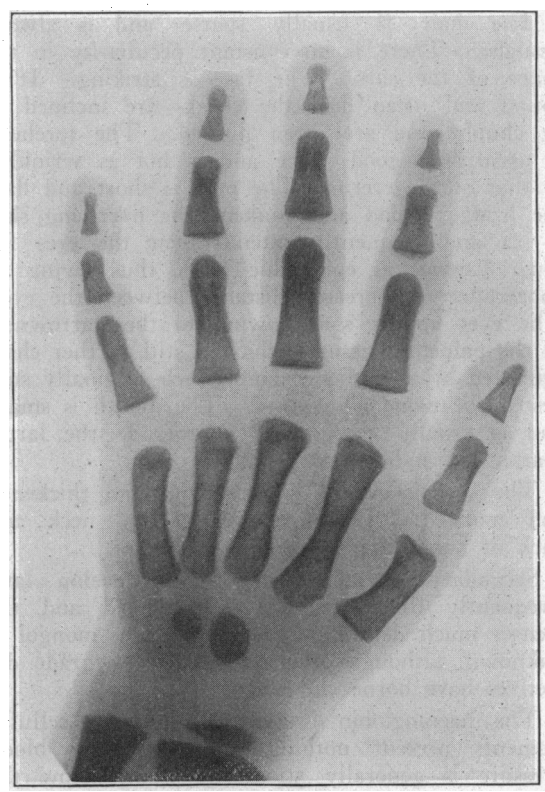

Fig. 4.
Radiogram of hand of Cretin, showing delayed ossification.

In a large number of cases the mother gives a history of menstrual disturbances, uterine displacements and pelvic operations, all antedating pregnancy. Is it not possible that some dysfunction of the internal secretion of the ovary and the related endocrine organs, analogous to an osteomalacia, may play an important part in the production of mongolism?

Jodicke and Dolega, in their study of metabolism in this disease, have found an increased combustion of sugar. This they believe is due to a hyperfunction of the pancreas, consequent to a hypofunction of the generative glands (ovaries and testes), which is shown clinically by their slowness and frequent lack of development.

THERAPY.

Thyroid extract has been given with the idea that there is a lack of function of the thyroid,

although the gland may be palpated in the majority of mongols. Brilliant results have been reported. There may, of course, be some lack of thyroid secretion in mongols, as there is in other individuals. The improvement must depend on the removal of the myxoedematous characteristics. Mongolism has not yet yielded to any glandular therapy.

Mongols respond to training, if taught by imitation. They are incapable of intellectual work involving calculation or imagination. The brighter children of this class may be trained to unskilled industrial work, particularly some phases of farming.

CONCLUSION.

The solution of the problem of mongolism lies in a better understanding of its metabolism and the interrelation of the endocrine glands, together with an intensive study of parental history.

REPORT OF TWO CASES OF AMAUROTIC FAMILY IDIOCY (TAY-SACHS' DISEASE).*

By CARL W. RAND, M. A., M. D.

From the Children's Hospital, Los Angeles, California.

Much has been written upon this disease since Warren Tay (1881) first described the unusual eye-picture in a mentally defective child that came under his observation. In his article entitled, "Symmetrical Changes in the Region of the Yellow Spot in Each Eye of an Infant," one reads; "In the region of the yellow spot of each eye there was a conspicuous, tolerably diffuse, large white spot, more or less circular in outline, and showing in its center a brownish-red fairly circular spot contrasting strongly with the white spot, and this white spot did not look like a hemorrhage or as if due to a pigment, but seemed a gap in the white patch through which one saw healthy structures." Later he described a similar condition in the eye-grounds of two other children in the same family.

In 1887 B. Sachs correlated the ophthalmoscopic findings with the neurological picture in a paper entitled, "Arrested Cerebral Development"; and in 1896 he further reported the nineteen cases then extant, giving the following symptom-complex as pathognomic of the disease, to which he gave the name "Amaurotic Family Idiocy":

1. Mental impairment in the first few months of life leading to absolute idiocy.

2. Paresis of the greater part of the body—flaccid or spastic in type.

3. Reflexes may be deficient or increased.

4. A diminution of the vision, terminating in absolute blindness (changes in the macula lutea and later an optic nerve atrohy).

5. Marasmus and a fatal termination as a rule about the second year.

6. The occurrence of the affection in several members of the same family.

7. Healthy at birth, remaining so to the third or fifth month; and occasionally—

8. Nystagmus.

9. Strabismus.

10. Hyperacuity of hearing.

11. Inordinate laughter was present in one case, and

12. Disturbances in deglutition were occasionally observed in others. It may also be added that the affection usually occurs in Russian Jews.

It seems superfluous at this time to review the literature which has become voluminous since 1896. Above one hundred cases have been reported to date, most of them in Jewish children. The pathology in brief consists of a peculiar degeneration of the ganglion cells of the entire cerebrum and cord, with a disappearance of the Nissel granules and a "ballooning" out of the diseased cell. Aside from the infantile type first described by Sachs, a juvenile form closely resembling it has been described by Vogt and others. Tay, Sachs, Carter, Vogt, Batten, Mayon, Higier, Ichikawa, Wandless, Dercum, Turner, Gordon, Kingdon, Hirsch, Cohen and Dixon, Schaffer, Spielmeyer, Mott, Bielschowski, Frey, Schob, Jacobi, Holden, Clairborne, Higier, Holmes, Gifford, Stewart Smith, R. M., Spiller and Wolfsohn have been the chief contributors.

The following case, No. 16878, was admitted to the Children's Hospital, Los Angeles, December 9, 1916. The patient, A. B., a baby girl, was admitted because of "helplessness and difficulty in swallowing." She was the youngest of four children, of intelligent Russian Jewish parents. The three other children died in infancy or early childhood,—two almost certainly of the same disease from which the patient died. The parents are both well and strong with a suspicious history of syphilis in the father dating back six years. The mother was married at seventeen, and her first baby, a boy, was born a year later. At ten weeks of age the child was stricken with "cholera infantum," and succumbed in a few days.

The second baby, a girl, was born two years later, when the mother was nineteen. She was a full term baby, birth normal, weight nine and a half pounds. For the first six months of life the child appeared normal in every respect, then began to lose weight, became weak, "paid no atten-

* Read before the Medical Society of the State of California, San Diego, Cal., April 19, 1917.